Chopin's illnesses

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Frédéric F Chopin (1810-1849) was an emotional and patriotic man who composed and played beautiful piano music and who died very young. He was also a very able crayon carciaturist but, regretfully, most of his drawings appear to be missing. His music is intellectually stimulating and often difficult. A reticent man by nature, Chopin left few informed accounts of his life. He was a diligent letter writer but some of his letters have not survived, either being accidentally destroyed by fires or even deliberately by his parents or friends if they contained politically sensitive material. He kept a very brief musical diary. Hence our knowledge of Chopin's life is based largely on some of his sketchy correspondence to his family and life-long friends: the accounts of his life as observed by George Sand, Liszt, Balzac, Hallé and a few other individuals who visited his Paris apartments between 1831-1849, either as musicians, pupils or literary scholars. Thus, some of the chronicles of fragments of his life are anecdotal essays or romanticized recollections which are impossible to verify and which may lead to speculative opinions.

A serendipitous discovery allowed me to read a great deal of Chopin's surviving letters to his parents and friends. They made fascinating reading and indicated to me that it was most unlikely that Chopin suffered from pulmonary tuberculosis[1] or cystic fibrosis[2] or even mitral stenosis[3] but another medical condition could explain his life-long malady. Four points emerged clearly from Chopin's correspondence:

(a) that he suffered from the same disease as his younger sister Emilia
(b) that his own life was going to be shortened as a result of it
(c) that the attending doctors did not know the nature of his disorder
(d) that his music became a symbol of his life's tragedy

The family history

Nicolas (a Frenchman by birth) and Justyna Chopin (Polish by birth) had four children Ludwika, Frédéric, Isabella and Emilia (Figure 1). Hardly any knowledge exists about the health of the respective grandparents. The father, Nicolas, was prone to develop infrequent respiratory tract infections, he became very ill on at least two documented occasions and survived to 70 years[4] (p 9). Justyna remained in good health and lived to 87 years - 'a neat, quiet, intelligent old lady'[5]. Ludwika, like her father, suffered from recurrent chest infections and died from a respiratory illness at the age of 47 years. Isabella led an uneventful life and died at 70 years. Emilia, the youngest daughter was described as a frail child, she suffered from periodic bouts of coughing, breathlessness and 'asthma'. At around 11 years of age she started to have haematemeses and died from a massive gastro-intestinal haemorrhage at the age of 14 years. By all the accounts of her very brief and painful life, Emilia, possessed a rare literary talent as manifested by a number of poems she wrote in German and Polish.

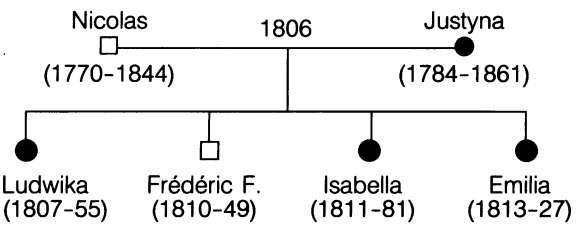

Figure 1. Chopin's family tree

Frédéric's medical history

There was an initial uncertainty about Frédéric's (and Emilia's but not Ludwika and Isabella's) date of birth and his christening was conducted within the first few weeks of life[6]. It is possible that both Frédéric and Emilia did not thrive well during the first months of life. As was the custom, all the children were born in the parents' bedroom in the presence of a 'midwife'. Frédéric was a delicate child, he tired easily on exertion and was not keen on strenuous physical activities. From his earliest years he was attended by doctors. During his early adolescence he developed intolerance to 'fatty foods' which resulted in prolonged episodes of diarrhoea and weight loss[7]. He experienced similar problems in 1838[8]. He was successfully treated with 'oats and honey' and often courses of belladonna which he continued to take through most of his life. 'I am quite well . . . as long as I avoid meats, sauces, soups, etc', he wrote in a letter to his parents from Berlin 27 September 1828.

He grew to about 170 cm (25th centile on the Tanner and Whitehouse charts) and was reported to be about 45 kg when 28 years of age (well below 3rd centile)[3] (p 140). The striking feature about his face was the lack of growth of facial hair and beard as noted by himself when 22 years of age, and others who described their observations while watching him during concerts. 'I have one side-whisker - the other won't simply grow . . .' (written winter 1832). Chopin was . . . 'whiskerless, beardless, fair of hair, and pale and thin of face . . . [9].'

He appeared to be sexually active in his early adult life (. . . 'a venereal disease', letter to Kumelski 18 April 1831; 'Me . . . a seducer?', letter to Tytus Wojciechowski 12 December 1831), occasionally got drunk and detested tobacco smoke which made him cough. Since he lived in the company of inveterate

Table 1. Chopin's doctors

Country	*Doctor*
1810-1830 Warsaw	W Malacz, F Girardot, F Roemer
1830-1848 Vienna	G Malfatti
1831-1848 Paris	A Hofman, A Molin, O Cauviere, J Matuszynski, P Gaubert, G Papet, J Coste
1848 London & Edinburgh	A Mallan, Sir James Clark
1848-1849 Paris	L Simon, J Fraenkel, J Koreff, P Louis, D Roth, J Creveilhier

cigar and pipe smokers such as George Sand, Liszt and his own father, Chopin undoubtedly became a chronic passive smoker for the best part of his life. We have no knowledge whether he ever fathered a child or whether he was capable of doing so. Certainly he mentions in his early letters about having children but the subject is not discussed either by George Sand in her memoirs or Chopin himself, despite their 11 years of cohabiting together.

As he entered adult life, Chopin continued to suffer from respiratory tract illnesses characterized by bouts of coughing; at times associated with productive cough and breathlessness, especially on exertion. Later on in life he had a number of episodes of haemoptysis which settled spontaneously. Despite clinical suspicion that his disease process might be due to consumption, none of his attending doctors in the last 10 years of his life was able to find any positive clinical findings: there was no digital clubbing, no lung cavity or any other lung disease.

> . . . the doctor examined him thoroughly and diagnosed no disease and no perceptible damage to the lungs, merely stressing that Chopin had a delicate chest . . .[10]
> Dr Papet . . . could find no 'lesions', no 'cavities' and no serious illness of any kind (letter, George Sand to Charlotte Marliani 8 March 1839)

During the early thirties he apparently became barrel-chested[3] (p 140).

Despite the negative clinical findings Chopin's health continued to deteriorate rapidly after the age of 30 years. The last 3 years of his life he spent 'vegetating', he suffered from episodes of haematemeses, severe headaches, neuralgia, disturbed sleep pattern and peripheral oedema. His creative musical output virtually ceased after 1845.

Autopsy result

Chopin requested for his body to be opened up so that the cause of his life-long malady could be determined. The autopsy was performed by Professor Jean Cruveilhier, a distinguished Paris pathologist (first Chair of Pathology in Paris) anatomist and physician and an authority on consumption. He issued the death certificate and wrote the medical report. Both these documents are missing. It is likely that they were destroyed in the Paris fires of 1871. It is well documented that Professor Cruveilhier had discussed the autopsy results with a number of individuals, namely Ludwika Chopin who was present at her brother's death, Adolf Guttmann, his favourite pupil, Wojciech Grzymala, his friend and Jayne Sterling, his former pupil and a generous benefactress. A review of their reports is fairly consistent

> The autopsy did nothing to disclose the cause of death . . . nevertheless he could not have survived . . . Diverse pathology . . . Enlarged heart . . . did not disclose pulmonary consumption. Lung changes of many years duration . . . a disease was not previously encountered.

and is also briefly summarized in an extract from a letter which Wojciech Grzymala wrote to Auguste Leo in October 1849[11].

> He gave instructions for his body to be opened, being convinced that medical science had never understood his disease and in fact it was found that the cause of death had been different from what was thought, but nevertheless he could not have lived.

Diagnostic possibilities

The course of Emilia and Frédéric's illnesses were characterized by chronic lung disease and gastrointestinal haemorrhages. A summary of the family's documented medical problems is given in Table 2. Emilia suffered from episodes of wheeziness and died following a massive haemorrhage most likely from portal hypertension or severe gastric erosion. Frédéric's lung disease dated back to childhood and the episodes of haemoptyses consisted of blood streaking purulent sputum - a picture of bronchiectasis or chronic bronchitis. The progress of his respiratory disease was marked by occasional respiratory tract infections, both upper and lower, from which he usually recovered almost fully and periods when his health appeared to be remarkably symptomfree. Emilia and Frédéric's medical conditions were very similar in their presentations and terminal outcomes, with the exception that in her case the disease process lasted about 10 years and in his case, well over 30 years.

The natural history of post primary pulmonary tuberculosis, cystic fibrosis or mitral stenosis is highly inconsistent with the illnesses suffered by Emilia and Frédéric. Assuming that both children had contracted tuberculosis, the source of infection remains a mystery. None of the immediate family members were

Table 2. Summary of documented medical problems of Chopin's family

Family member	*Medical condition*
Nicolas	Chest infections; died at 74 years
Ludwika	Chest infections; death at 47 years from chest infection
Frédéric	Frail
	Dietary problems
	Recurrent respiratory problems - upper and lower
	Weight loss and weakness
	Haematemeses
	Haemoptyses
	Facial hair loss
	Peripheral oedema
	Barrel-chest
	Bone pains
	Terminal respiratory failure; at age of 39
Emilia	Frail
	'Asthma'
	Haematemeses
	Terminal massive gastric haemorrhage at age of 14 years

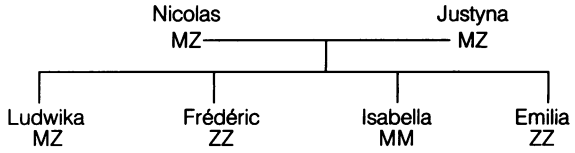

Figure 2. Hypothetical Pi phenotypes for the Chopin family

known to have died of consumption or to have had typical features of tuberculosis disease. A friend was known to have had consumption and was cared for by Frédéric but this occurred in the last years of his life when his disease was already well advanced. The spread of tuberculosis from Emilia to Frédéric, as suggested by some[1] is highly unlikely. Had Emilia suffered from miliary tuberculosis or a direct progression of a primary lesion, her clinical symptoms would have been quite dramatic, and the death would have occurred rapidly within 1-5 years. However, Emilia did not demonstrate any impressive symptomatology for some years ('asthma-like' symptoms only) until she started to have bouts of haematemeses at about 11 years of age which ultimately were responsible for her death 3 years later. Frédéric during the early stages of his life had had symptoms suggestive of active tuberculous lung disease. However, none of the physicians were able to diagnose pulmonary tuberculosis convincingly and Professor Cruveilhier's autopsy failed to confirm pulmonary tuberculosis although he did detect 'a disease not previously encountered'. Death due to cystic fibrosis lung disease in the pre-antibiotic era was uniformly fatal during the first 5-10 years of life. It would have been quite remarkable for Frédéric to survive active lung disease to 39 years of age and to demonstrate such a protracted manifestation of symptoms and signs, many of which were not typical of cystic fibrosis.

The unifying hypothesis which explains best Emilia and Frédéric's illness points to α1 antitrypsin deficiency (α1 AT) as the explanation. This genetic deficiency of the serine protease inhibitor α1 AT is accompanied by early onset of degenerative lung disease and liver damage which is responsible for at least 25% of deaths from juvenile cirrhosis[12-14]. The name of α1 AT was originally suggested by Schultze *et al.* (1962)[15] for the protein his group described in 1955. The history of Protease Inhibitor (Pi) system dates to the observations of Lovell and Eriksson (1963)[12] that in the sera of some patients α1 globulin band was absent on paper or agarose electrophoresis. This deficiency was specifically associated with the early onset of emphysema, especially in smokers but it may be also associated with bronchiectasis and chronic bronchitis. Some patients developed cirrhosis of the liver. A large number of alleles of α1 AT gene have been identified. The most important type deficiency is associated with the ZZ phenotype[13]. The hypothetical family Chopin Pi phenotypes is given in Figure 2.

In parts of Europe, one in 10 individuals are carriers of the deficiency gene and about one in 1000 has a genotype PiZZ which predisposes them to the development of both lung and liver disease. About 4% of Europeans are heterozygotes for the Z variant[13] (PiMZ). The phenotype data for parts of France are available[14] but not for Poland. The heterozygote adults type MZ have three times the risk of normal population for developing respiratory disease[16]. In families with an affected older sibling (Frédéric) the chances of developing juvenile cirrhosis in subsequent affected children (Emilia) is increased considerably[17]. The natural history and life expectancy in a large group of patients with α1 AT, PiZZ is shown in Figure 3 and Table 3.

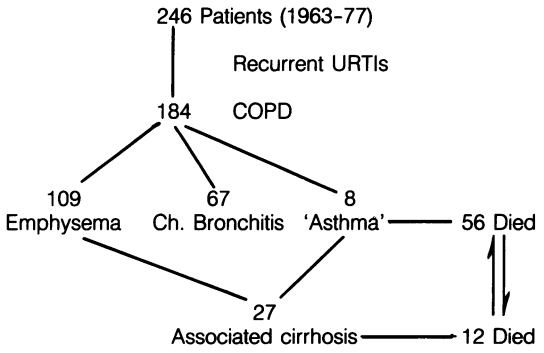

Figure 3. Natural history and life expectancy in α_1 antitrypsin deficiency, PiZZ. Larsson C, Acta Med Scand 1978;**204**:345

Conclusions

Frédéric and Emilia suffered from α1 AT deficiency. Emilia died of bleeding from oesophageal varices due to juvenile cirrhosis of the liver. Frédéric developed bronchiectasis early in life and in the last 10-16 years of his life showed features of progressive liver damage, e.g. hypoproteinaemia, feminized features, gastrointestinal bleeding. The cause of his death was most likely due to acute respiratory failure as a result of obstructive lung disease and cirrhosis of the liver. Would Chopin have composed the exciting piano music had his health been perfectly normal? I doubt it. In my view, his third piano sonata in B minor, one of his greatest works, written in the summer of 1844 when his health was rapidly failing, proclaims in the closing bars his ultimate victory over his life's tragedy. As Liszt perceptively observed, Chopin

> possessed a great physical strength . . . and hardly allowed one to suspect the secret convulsions which agitated him[18].

Table 3. The natural history of α1 antitrypsin deficiency, type PiZZ (from Larsson C, Acta Med Scand 1978;**345**. *Svager T,* N Engl J Med 1976;1316. *Psacharopoulos HT,* et al. Arch Dis Child 1983;**58**:882*)*

Infant	*Child*	*Adult*
Neonatal hepatitis	Fulminating juvenile cirrhosis	Slowly progressive liver damage
'Failure to thrive'	Asthma-like symptoms	Respiratory infections
Recurrent wheeze	Respiratory infections	Panacinar emphysema

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Based on the Presidential Address read to Section of Paediatrics, 22 March 1994

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Letters to the Editor

If Cytokines be the Food of Love[1]

Johann Wolfgang Goethe, the 'Prince of German poets', inadvertently expressed the physiology of love and also, perhaps, the condition of cyclothymia in his drama *Egmont.* In the third Act (Scene 2), Klaerchen, Count Egmont's sweetheart, sings a little song, in which occur the lines:

'Himmelhock jauchzend,	Jubilant to high Heaven
Zum Tode betruebt -	Distressed to Death,
Gluecklich allein	Happy only
Ist die seele, die liebt.	Is the Soul in Love.

Goethe wrote this in 1787!

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Beethoven's nephropathy and death

Dr T J Palferman's belated clever, attempts to discredit my diagnosis of renal papillary necrosis (April 1994 *JRSM*, p 247) fail to convince. The cornerstone of this dispute is the consistency of the calcareous concretions rather than semantics.

Palferman makes light of the pathologist Wagner's choice of the adjective 'calcareous' in preference to the noun 'calculus', yet Dr Wagner's description was clear and unequivocal.

When Dr Wagner cut sections through Beethoven's kidneys he was surprised to find unusual lesions in every single calyx. He carefully chose the Latin words 'concremento calcareo'. There is no doubt that he was describing chalky concretions, of the consistency of carbonate of lime, since he went on to state that he was able to cut through them to produce an appearance 'like a pea which had been cut across the middle'.

There is, therefore, no question that these lesions were not stony hard renal calculi, which Dr Wagner would have been unable to cut across the middle!

Palferman has erred in failing to consult the original sources. Paradoxically in his articles he describes the lesions as 'chalky calyceal concretions the size of split peas', but he fails to cite his source. In doing so he inadvertently failed to recognize the distinction from renal calculi.

Nor did Palferman mention the vital clue that concretions were present in every calyx. When, as occurs especially in diabetic cases, all the papillae of both kidneys are effected, the diagnosis of renal papillary necrosis is not in doubt, since no other condition can be responsible.

Yet Palferman insists that 'the likelihood remains of nephrolithiasis secondary to sarcoid induced hypercalciuria and hypercalcaemia'. This is impossible since the changes described by Dr Wagner are incompatible with nephrocalcinosis, where the calcifications are deposited in the tubules, especially of the pyramids, or in the parenchyma. While at times small calculi may form or be extruded into the renal pelvis, it must be emphasized that the lesions found in Beethoven's kidneys are quite dissimilar to the nephrolithiasis of sarcoidosis!

Beethoven's history of chills and abdominal pain is consistent with though not diagnostic of renal papillary necrosis. In commenting on Beethoven's medical history Palferman trips himself over unwittingly when misquoting me.

After having stated correctly that I had proposed that a terminal diabetes was responsible for the renal papillary necrosis, he then states that 'diabetes was surely not present all this time (1792-1801-1816)'.

Palferman misquotes me again on my 'insistence that the sloughed papillae caused ureteric colic'.

I stated that 'There were clearly insufficient clinical data for us to be certain of the diagnosis . . . it might have been a renal colic'.

Palferman errs yet again when he states that 'loss of pancreatic exocrine function . . . leading to steatorrhoea and malabsorption was absent in Beethoven'. The composer's pancreatic exocrine function was never estimated, nor has any record of description of his stool survived.

It is not possible for me to comment further here on Dr Wawruch's report or the abdominal symptoms. A detailed discussion is given in my forthcoming book on Beethoven's health.

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